Topics in Primary Care Medicine

Dupuytren's Disease

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"Topics in Primary Care Medicine" presents articles on common diagnostic or therapeutic problems encountered in primary care practice. Physicians interested in contributing to the series are encouraged to contact the series' editors.

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Dupuytren's disease is a fibrotic thickening of the palmar fascia initially manifested as painless nodules of one or both hands and subsequently by permanent contractures of one or more fingers into the palm. Every primary care physician sees Dupuytren's disease because it is the most common affliction of the hand, affecting 25% of persons of Celtic ancestry and as many as 5% of the general population. Interesting associations with other localized fibrotic disorders such as knuckle pads (Garrod's nodules), Peyronie's disease, and plantar fibrosis (Ledderhose's disease) have been recognized. Primary care physicians should be prepared to recognize the various stages of the disease and to make an appropriate referral before disabling and often irreversible contracture occurs.

Since Baron Guillaume Dupuytren's description of a peculiar retraction of the fingers into the palm in 1831, the disease that bears his name has come under intense scrutiny and yet remains an enigma. First noted by Plater in 1680, referred to in lectures by Henry Cline in 1808, and related to hypertrophy of the palmar fascia by Sir Astley Cooper in 1822, the strange affliction was popularized by Dupuytren in 1831 in his classic account of permanent retraction of the fingers in the first chapter of his work, *Leçons Orales de clinique chirurgicale faites a l'Hotel-Dieu de Paris.* Dupuytren described the first known surgical correction, which entailed making transverse incisions through the palmar aponeurosis and through the contracture at the proximal interphalangeal joints. After two months of splinting, the patient regained normal function. In the same work, Dupuytren gave a description of symptoms and findings that is still useful. He further stated that the palmar aponeurosis was the cause of the retraction, discounted influences of the tendons and skin, and hypothesized a traumatic etiology.

Slightly more than a year later, Dr G. Gayrand at the Hotel-Dieu at Aix presented new findings to the medical academy in Paris in April of 1834. He noted that the skin and subcutaneous tissues were actively involved but with little involvement of the palmar aponeurosis, and with fibrous cords passing from the aponeurosis to the flexor tendon sheaths. He questioned a traumatic cause, noting bilateral involvement in a patient who had not performed manual labor. Finally, he questioned Dupuytren's technique, calling for a longitudinal incision with excision of the contractile band. Currently the controversy has been carried into the courts as members of the legal profession now debate the same issues to determine financial compensation to workers of various occupations so afflicted.

Anatomy

The palmar fascia is composed of a superficial and deep layer, with anchoring fibers passing to the palmar skin superficially and to the interosseous fascia deeply (Figure 1). The superficial palmar fascia (of the palmar aponeurosis) is a broad, triangular sheet with its apex at the wrist and its distal portions dividing into longitudinal slips as it passes toward the fingers. Most of the distal fibers pass upward to the skin, forming a strong attachment between the palmar fascia and the skin, most prominently at the distal palmar crease. Other fibers run transversely to form the interdigital ligaments of the finger webs or deeply to join or interdigitate with fibers ascending into the finger. The deep palmar fascia is predominantly transverse and thickened centrally, passing from the transverse carpal ligament to the four central digits. Passing downward from the deep fascia are fibrous slips that form eight tunnels for the lumbricals and the flexor tendons of the four central fingers. Thus, the palmar fascia fixes the skin during gripping and twisting maneuvers, impedes hyperextension of the fingers, and provides functional stability to the flexor mechanism. Dupuytren's disease variably involves portions of the entire apparatus.

Histology

The lesions of Dupuytren's disease resemble normal scar tissue with fibroblastic proliferation and collagen accumulation. The nodules are more cellular than the longitudinal bands, with numerous fibrocytes arranged in a concentric pattern. The bands are devoid of cells and are composed of wide sheaths of collagen fibers. The most histologically active lesion appears to be the subcutaneous nodule, which is probably the initiating site. Fibrosis may extend to the

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dermis, accounting for the skin contractures and dimples. The histologic appearance of the knuckle pads and the plantar nodules is similar.

Three histologic phases occur in sequence, much as in the formation of normal granulation tissue. The early proliferative phase is characterized by fibroblastic proliferation. The appearance of myofibroblasts and type III collagen marks the involutional phase, and the residual phase is marked by the appearance of fibrocytes and type I collagen until an acellular dense band is formed. The presence of myofibroblasts in nodules or fibroblasts with microtubular structures in the fascia of resected specimens is predictive of recurrence. Increased numbers of myofibroblasts and type III collagen can also be found in grossly normal aponeurotic tissue in Dupuytren's disease, serving as sites for postoperative recurrences.

Etiology

The initiating event that propagates the cascade of fibrosis remains unknown. Since Dupuytren's time, the relative contribution of trauma or microtrauma has been debated. Dupuytren's disease is not related to occupation, nor is it more prevalent in those performing manual labor. It involves the nondominant as often as the dominant hand and the ulnar rather than the radial aspect. No suitable explanation has been given for the often-noted association with alcoholism, epilepsy, or diabetes mellitus. Specific hormone receptors have not been found in the fibrotic band, nor has research uncovered the nature of the predisposition in men. A dominant hereditary pattern appears to be present in those of Celtic ancestry. A positive family history is reported by 25% of unselected patients, with large kindred investigations finding aspects of the disease in 68% of first-degree relatives of affected patients, suggesting an autosomal-dominant pattern of inheritance. Antibody to denatured type II collagen is found more commonly in those of the HLA-DR4 genotype. Thus, there appears to be a genetic predisposition that sensitizes a person to form a contracture after a yet-unknown stimulus.

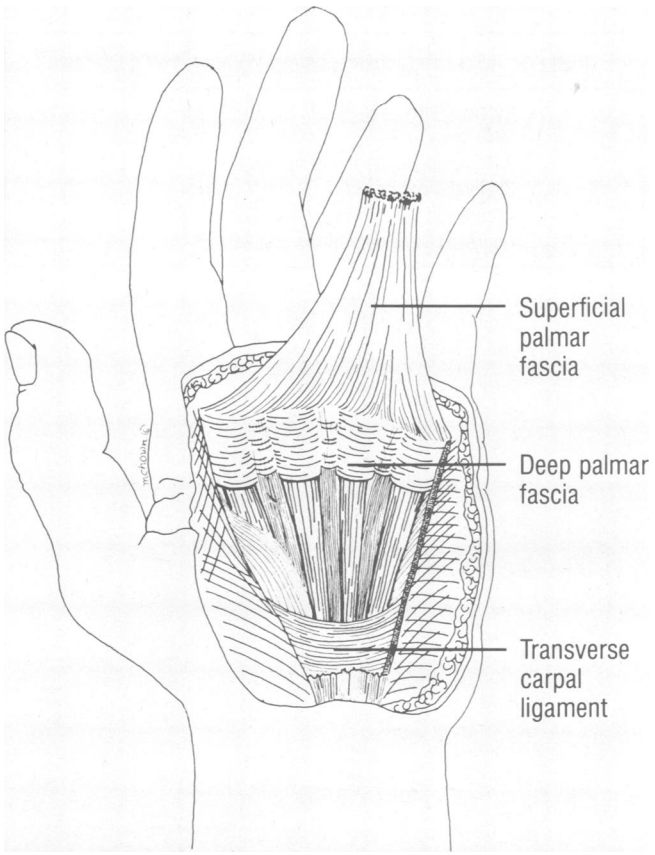

Figure 1.—The deep transverse and superficial longitudinal palmar fasciae are depicted. Note the tunnels formed for the passage of the lumbricals and flexor tendons. Dupuytren's disease involves components of the entire apparatus.

Incidence

The prevalence of Dupuytren's disease is greatly influenced by the population studied. It is found almost exclusively in persons of Celtic ancestry. Its geographic distribution reflects patterns of Celtic migration during the 5th century to Britain and western Europe. Subsequent selective immigration to Australia and the eastern coast of North America accounts for the high prevalence rates in these areas. Dupuytren's disease is extremely uncommon in persons of African or Asian descent. It is present in 4% to 5% of unselected populations, but in Melbourne, for example, the disease affects 20% of the population aged 60 and older and 40% of those aged 80 and older. In the British Isles, 25% of men older than 65 are so afflicted, and in France Dupuytren's disease was found in 12% of nonalcoholic patients averaging 54 years of age. Men are affected twice as often as women before age 40, but the difference lessens thereafter, although in men the disease more commonly progresses to flexion deformities. The disease is rare before age 25, typically occurring after age 40.

Associations with epilepsy, alcoholism, diabetes mellitus, and chronic pulmonary tuberculosis have been noted, although no association with epilepsy was reported until after the introduction of anticonvulsants in 1940. Hueston cites a 43% incidence in men with epilepsy younger than 40 years, a 44% incidence in alcoholic patients, and a 42% incidence in patients with chronic pulmonary tuberculosis. Prospective studies using univariate and logistic regression methods on patients admitted to hospitals have shown a significant relationship between age, total alcohol consumption, and Dupuytren's disease, with the severity of disease related only to the amount of alcohol consumed. No relationship to chronic liver disease or manual labor was noted.

Clinical Aspects

Dupuytren's disease is usually bilateral (65%) but often not symmetric. The palm is involved in almost all cases (95%), with the ring or small finger involved in as many as 75% of patients. The middle finger is affected in as many as 75% of patients. The index finger is involved least frequently (13%), and the thumb and long fingers are affected in about a third of cases.

At first a small painless nodule may appear at the distal palmar crease opposite the affected finger. The skin generally becomes fixed to the underlying fascia with a characteristic dimpling. This usually presents no more than an inconvenience to the patient and is easily ignored. Pain at this stage should suggest another cause. The fibrotic process may not progress further, as is often the case in women, or may begin to cause flexion contractures at the metacarpophalangeal joint of the ring and subsequently the adjoining fingers. Later the proximal interphalangeal joint may become affected as well.

The transition from palmar nodules to flexion contracture may occur rapidly over the space of a year or more slowly over as long as two decades. The proximal interphalangeal joint may then become involved, flexing the finger into the palm and completely inhibiting normal grip. The hooklike contracture may predispose to injuries because the contracted fingers catch on machinery or rails (Figure 2). The skin macerates and becomes secondarily infected at the level of the flexion creases or interdigitally.

The association of Dupuytren's disease with knuckle pads, plantar fibrotic nodules, and Peyronie's disease has been called the "Dupuytren's diathesis." Of patients so affected, 44% show tender nodules over the dorsum of the proximal interphalangeal joints, preventing the wearing of rings. The nodules (Garrod's knuckle pads) appear earlier than the palmar nodules and often regress as the other manifestations of the disease progress. Plantar nodules are seen in 18% of patients, although they also occur at an earlier age and can become large and painful. Histologically they can be confused with fibrosarcoma, leading occasionally to inappropriate amputation. The nodules usually regress spontaneously, often followed by further recurrences. Attempted surgical removal usually leads to additional disability. In 3% of patients with Dupuytren's disease, signs of Peyronie's disease will develop. Peyronie's disease is a fibrotic disorder of the penile shaft leading to anterior angulation. A familial transmission of Peyronie's disease has been reported, with 78% of affected persons also having Dupuytren's disease. The use of β-adrenergic blockers has been associated with Peyronie's disease, although no such association has been found in Dupuytren's disease. The Dupuytren's diathesis also includes shoulder stiffness, fascial fibromatosis, a loss of joint mobility, a loss of thoracic and lumbar spine motion, and diffuse fibrosis of the shoulder, hip, anterior leg, or antecubital space.

The diagnosis is usually straightforward when physicians are presented with a thickened palmar aponeurosis and associated finger contracture. Occasionally, however, other conditions mimic the contractures caused by Dupuytren's disease (Table 1). Traumatic linear midline scars can cause

TABLE 1.—*Differential Diagnosis of Dupuytren's Disease*

Differential Diagnosis	*Differentiating Factor*
Congenital flexion deformity	Appropriate history
Posttraumatic scar	History of trauma or burn
Flexion contraction due to immobilization	Appropriate history
Volkmann's ischemic contracture	Wrist and PIP joint flexed
Primary joint contracture	Palm unaffected, painful joint, associated systemic disease evident, history of trauma
Postinflammatory flexor tendon contraction	Trigger finger
Fibrosarcoma of the palm	Does not progress to contracture, painful
Plantar fasciitis	Associated with painful arthritis and systemic malignancy

PIP=proximal interphalangeal

TABLE 2.—*Poor Prognostic Factors in Dupuytren's Disease*

- Positive family history
- Male sex
- Associated alcoholism or epilepsy
- Bilateral involvement
- Associated knuckle pads or plantar nodules
- Aggressive behavior of the contraction

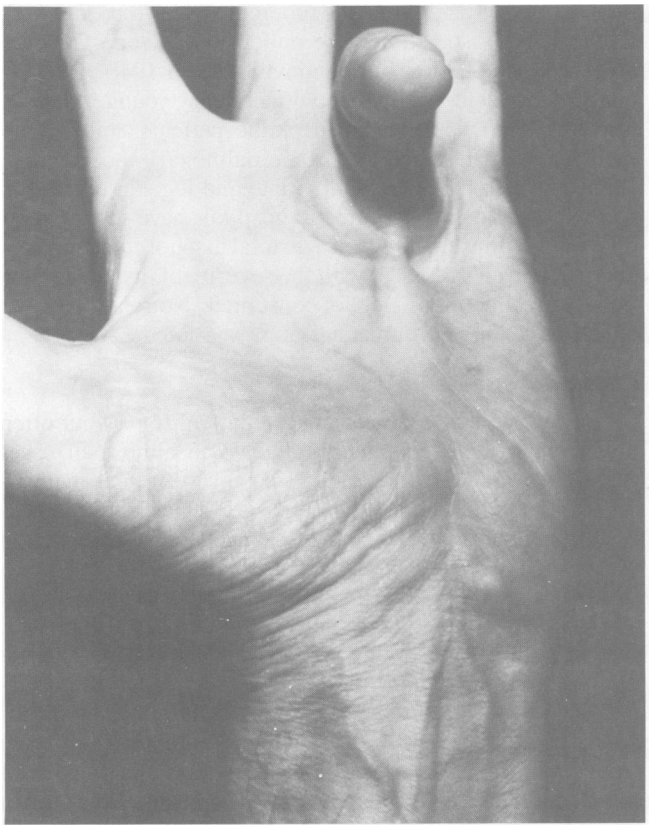

Figure 2.—Dupuytren's disease is shown involving the ring finger. Note the thick contractile band, skin dimpling, and the hooklike contracture that predisposes to an accidental attachment to machinery and railings.

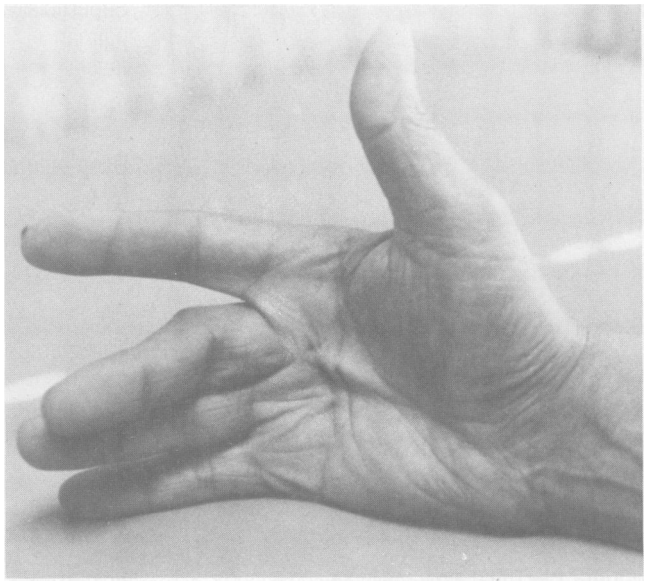

Figure 3.—Dupuytren's disease is shown involving the middle finger. Note the slight contracture of the proximal interphalangeal joint—an indication for immediate surgical correction.

contractures of the digital joints. In contrast to the contractures of Dupuytren's disease, in which the contracting band is dermal or subcutaneous, linear traumatic scars are limited to the superficial layers of skin. Joint contractures after trauma or arthritis can be differentiated by history and x-ray films. Although a damaged flexor tendon may result in a flexion deformity, there is no palpable subcutaneous band. A "dropped finger" of an extensor tendon injury can be corrected passively, whereas a contracted finger in Dupuytren's disease is fixed. A long-standing trigger-finger deformity can cause locking of the digit, but the locked tendon can be released by sharply flexing the finger at the middle interphalangeal joint while passively extending the digit. Knuckle pads and plantar nodules should alert the physician to the presence of Dupuytren's disease as well.

Prognosis

Milford has defined six factors that indicate a poor prognosis (Table 2). Contractures involving the central longitudinal pretendinous band respond well to surgical treatment. Involvement of the deep transverse intermetacarpal ligament, however, with its attachment to the joint capsule or the proximal interphalangeal joint and the associated entrapment of the digital nerve, makes repair hazardous and often ineffective. Contractures of the metacarpophalangeal joints are more easily repaired than are those of the proximal interphalangeal joints, with the latter often impossible to repair in neglected cases.

Treatment

No satisfactory treatment yet exists for Dupuytren's disease. Nonsurgical treatments such as local corticosteroid injections, ultrasound, topical ointments, and vitamins have been tried without success. The traditional method of sitting on one's hands, palmar side down, is probably as effective as most treatments in the early stages. Treatment strategies must be tempered by the extreme variability of disease presentation and the rate of progression. Several points are noteworthy, however. Simple nodular disease of the palm need not be approached surgically. Patients with nodules may have no further progression and may remain asymptomatic indefinitely. Minimal flexion contractures of the metacarpophalangeal joints can be stable and nondisabling and can be repaired successfully later. Even minimal involvement of the proximal interphalangeal joint should be surgically repaired immediately (Figure 3) because permanent contracture not amenable to later surgical repair is likely to occur. In all cases, cautious observation is recommended with careful instructions to patients to return immediately if any interphalangeal involvement develops.

The choice of surgical therapy depends greatly on the degree of involvement, a patient's age and general physical status, and whether the operation is being done for a recurrence. Each approach necessitates considerable surgical skill and experience. In an elderly or infirm patient, a limited subcutaneous fasciectomy can be done that is palliative, but it is associated with a 72% recurrence rate. A partial selective fasciectomy suffices when only one or two ulnar fingers are involved. This limited operation is associated with a low complication rate, and, although recurrence is again frequent, only 15% need another operation. Both diseased and grossly normal palmar fasciae are removed by complete (radical) fasciectomy. This operation has generally been abandoned because of an unacceptable complication rate. Occasionally a limited fasciectomy can be combined with an extensive fasciectomy of any involved finger. Fasciectomy with skin grafting offers an excellent choice for a patient with a poor prognosis or in those with recurrence. The complication rate is low, and the procedure offers the lowest recurrence rate.

Complication rates generally reflect the skill and experience of the surgeon and include hematoma, skin necrosis, infection, digital nerve transection, sympathetic dystrophy, and recurrence. Only occasionally is a digit lost from a surgical mishap. Recurrence rates continue to be about 20%, reflecting the continued progression of the fibrotic process despite temporary surgical correction.

Summary

Dupuytren's disease is an extremely common malady, affecting as many as 3% of the general population. Presenting features are variable and include simple asymptomatic palmar nodules or refractory contractures of the interphalangeal joints. Substantial associations with knuckle pads, plantar nodules, and Peyronie's disease are noteworthy. Although a strong familial tendency is present, the precise pathologic mechanism is unknown. Treatment is frequently unnecessary, but when indicated it includes a variety of surgical alternatives. An appropriately timed referral to a surgical specialist before irreversible contracture of the interphalangeal joints can prevent a permanent loss of function. When surgical intervention is not elected, careful and regular follow-up is necessary to detect early joint contracture.

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